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Case Report

Retroperitoneal hematoma complicated with colocutaneous fistula in a case of severe hemophilia B*

Haydar Zengin[1], Melda Cömert[1], Tuba Bulduk[1], Rahman Şenocak[2]

[1]Department of Hematology, Gülhane Training and Research Hospital, University of Health Sciences, Ankara, Turkiye
[2]Department of General Surgery, Gülhane Training and Research Hospital, University of Health Sciences, Ankara, Turkiye

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ABSTRACT

Retroperitoneal hematoma rarely occurs in hemophilia B cases caused by factor IX deficiency. It typically arises as a result of trauma or surgical procedures, though spontaneous cases are rare. We present our patient who developed spontaneous retroperitoneal hematoma and was subsequently complicated by abscess, coloenteric fistula, osteomyelitis and arteria lumbalis bleeding. Surgical intervention was avoided for a long time due to difficulty of hemophilia B management for surgery. Therefore, the complication became severe and life-threatening. The patient was treated with successful and long-lasting surgical interventions, intravenous antibiotics and factor IX replacements. To the best of our knowledge, the case we present is the first hemophilia patient in the literature complicated by colocutaneous fistula.

Keywords: Hemophilia-B, retroperitoneal hematoma, colocutaneous fistula

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INTRODUCTION

Hemophilia B is a rare bleeding disorder that is transmitted in an X-linked manner and occurs in approximately 1 in 25,000 male births.[1] The severity of bleeding in hemophilia B is inversely proportional to the factor level. The most common bleeding sites are intra-articular and intra-muscular. Retroperitoneal bleeding is extremely rare in hemophilia B, with its exact frequency remaining undetermined due to the limited data available from case reports.[2,3]

Although there have been significant advances in the treatment of hemophilia, most of the patients around the world still cannot access these treatments due to social and economic problems. For this reason, serious complications can still be seen in undeveloped and developing countries.

CASE

A 37-year-old male patient was diagnosed with hemophilia-B at the age of 13. He could not walk at the age of 12 because of arthropathy in both knees due to the delay in diagnosis. The patient had an operation at an external center 3 years ago due to retroperitoneal hematoma and then received intensive factor therapy due to recurrent bleedings, developed multiple skin fistulas and then colon fistulas at the operation site and around it. Radical procedures could not be performed because of the anatomical location of the bleeding and the complications with infection. Intensive antibiotic treatment had to be applied during this process. The patient was hospitalized with the existing retroperitoneal hematoma turning into a fistulizing abscess on the colon+lumbar region skin. The lesions at the time of admission and the healing findings are shared in **Figure 1** after the informed consent form was obtained.

Figure 1. a) Discharged fistula lesions at first admission to the hospital, b) Clean skin fistulas after the surgery, c) Ileostomy

The patient had colonic contents leaking from the fistula mouths; there was a heavy anaerobic odor emanating from the wound areas and putting a strain on his social life. His laboratory findings at the time of admission were as follows: Hgb: 6.4 gr/dl, WBC: 6.3x10^9/L, Plt: 207x10^9/L, aPTT: 49.8 sn, INR: 1.2, CRP: 190 mg/L. Wound cultures yielded *E. coli*, *Morganella morganii*, *Proteus mirabilis*, and blood culture yielded *Psychrobacter*. was given Intravenous antibiotics was given. Abdominal tomography showed an image that started from the posterior neighborhood of the left kidney and spleen that extended to the pelvis, opened to the skin, contained air densities and contrast material extravasation suggesting active bleeding (originating from the left a. lumbalis and the

Corresponding Author: Haydar Zengin, hfab65@gmail.com

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aorta), could not be distinguished from the bowel loops, and eroded the left 12th rib, L4 and L5 vertebral processes, and left iliac bone. The radiological image is shown in **Figure 2**. The general surgery team performed extensive debridement of the complex hematoma and abscess, which extended into the abdominal and retroperitoneal regions. Ileostomy was performed, colon and skin fistulas were repaired. During the hospitalization, which lasted approximately 7 months, the patient underwent numerous surgeries for wound site revision. All skin fistulas closed; the source of infections was eliminated. With the use of effective factor IX replecaments both before and after the operation, the patient did not experience any bleeding and hemostatic problems.

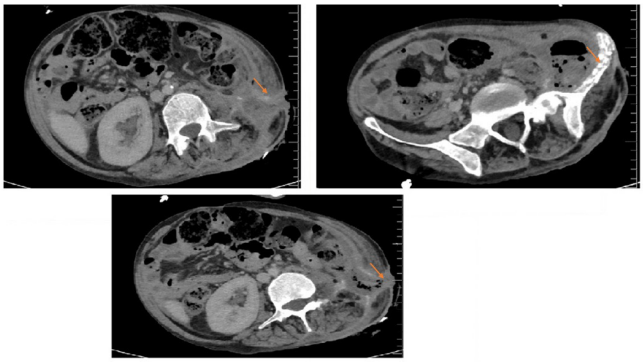

Figure 2. Computerized tomography images showing the patient's colocutaneous fistula and the deterioration of the structure of the iliac bone

DISCUSSION

Fistula is defined as an abnormal connection between two body structures that should not be connected.[5] Colocutaneous fistulas mostly develop as a complication of Crohn's disease, diverticulitis or surgical intervention.[5-7] Our patient also underwent an intervention for hematoma drainage 3 years ago.

It has been stated in the literature that it is important for enterocutaneous fistula treatment to be performed in a center with serious experience and to use a multidisciplinary approach.[9] Similarly, retroperitoneal hematoma is an important condition with high mortality, where early diagnosis and correct treatment are important.[8] Intervention in a patient with severe hemophilia who develops two serious complications, retroperitoneal hematoma and enterocutaneous fistula, can be frightening. However, these conditions with high morbidity and mortality risks can be managed by performing effective factor replacement at an early stage. In late cases, a long process requiring patience is required with the cooperation of hematology, general surgery and infectious diseases. The shared case has identified this situation.

Surgeons should be encouraged to intervene early in life-threatening and life-disrupting complications such as colocutaneous fistula in hemophilia patients. The most important task in this regard falls to internal medicine specialists and hematologists.

CONCLUSION

Despite the advances in hemophilia treatment in recent years, there may still be complicated cases. It is more common in undeveloped and developing countries where access to treatment is limited. Early intervention in complications that develop in hemophilia cases will increase the chance of success. When surgery is necessary in hemophilia patients, it is necessary not to hesitate to intervene. Good relationship and multidisciplinary approach that encourages courage should be established between the hematologist and the surgeon.

ETHICAL DECLARATIONS

Informed Consent

The patient signed and free and informed consent form.

Referee Evaluation Process

Externally peer-reviewed.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

Financial Disclosure

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Author Contributions

All of the authors declare that they have all participated in the design, execution, and analysis of the paper, and that they have approved the final version.

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